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Global Transient Amnesia as the Predominant Symptom of Cardiac Myxoma: A Case Report

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Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
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Patient: Female, 67-year-old
Final Diagnosis: Cardiac myxoma
Symptoms: Transient amnesia • gait imbalance • dizziness
Medication: —
Clinical Procedure: Resection of cardiac tumor
Specialty: Cardiology • Neurology

Objective: Unusual clinical course

Background: The most common neurological symptoms from cardiac myxoma-induced stroke include territories of middle cerebral arteries, rendering posterior stroke less common. Although transient global amnesia usually has a benign prognosis, amnesia in the setting of concerning cerebellar symptoms should raise the suspicion for posterior circulation involvement. These benign-appearing symptoms can be manifestations of an acute cerebrovascular accident (CVA). This unusual presentation can delay workup for underlying pathology.

Case Report: A 67-year-old woman presented to the local emergency department after an episode of global amnesia that lasted about 15 minutes and was associated with some dizziness. The patient also reported a history of chronic disequilibrium. The head CT scan was negative for any acute findings. A follow-up MRI of the brain demonstrated acute small lacunar infarcts within the left cerebellum and right parietal lobe. An echocardiogram was performed due to concern for the cardioembolic source, which revealed left atrial myxoma. She was transferred to a tertiary center for immediate surgical intervention due to the high risk of embolization associated with the condition. The patient subsequently underwent successful surgical excision of the lesion.

Conclusions: Cardiac myxoma, although a rare cause of posterior stroke, needs prompt intervention as it is associated with a high risk of systemic embolization, including recurrent CVA. Transient global amnesia is an atypical presentation of cardiac myxoma that can easily be overlooked, delaying timely diagnosis and prompt intervention. Early recognition and surgical resection are crucial to prevent potentially life-threatening consequences.

Keywords: Myxoma • Brain Infarction • Ischemic Attack, Transient • Amnesia, Transient Global

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/938221

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Background

Embolization from cardiac myxoma is not an uncommon phenomenon. However, it is quite uncommon for transient amnesia to be the predominant symptom from embolization of cardiac myxoma. Clinical manifestations may vary greatly, from obstruction of blood flow, tempering valvular structures, to embolization. In most cases, the tumor releases fragments or thrombi into the systemic circulation, leading to embolization. The most serious and potentially life-threatening complication of cardiac myxoma is cerebrovascular accident (CVA) secondary to embolization. The most common presentations of cardiac myxoma-induced stroke include hypoesthesia, hemiparesis, and facial paresis due to cerebral infarction [1]. Here, we present a unique case in which nonspecific dizziness and transient global amnesia were the predominant presenting symptoms in our patient, who was subsequently found to have multifocal infarct involving the cerebellum. Atypical presentation of this nature can undermine suspicion for underlying stroke and thus delay prompt diagnosis and treatment [1,2].

Case Report

A 67-year-old woman initially presented to the Emergency Department (ED) following an episode of transient memory loss with associated dizziness. Her medical comorbidities included history of breast cancer status after bilateral mastectomy and curative chemotherapy without recurrence, peripheral neuropathy from chemotherapy, hypertension, hyperlipidemia, type 2 diabetes, heart failure with mid-range ejection of 45-50%, morbid obesity (body mass index 45), mild obstructive sleep apnea, and asthma. The patient reported that her episode of global amnesia lasted approximately 15 minutes and was preceded by mild lightheadedness. Upon further questioning, she reported a long-term history of difficulty with gait instability and tendency to veer toward the left side, which was intermittent. In addition, she also noticed occasional difficulty with balance, not related to positional change. She denied having headaches or change in vision and hearing, had no trouble swallowing or pain with swelling, and no new numbness or weakness or tingling. All laboratory tests in the ED were within normal limits. A head computed tomography (CT) scan done in the ED was negative for acute findings. Magnetic resonance imaging (MRI) of the brain showed acute lacunar infarcts present within the left cerebellar hemisphere, measuring 7×4 mm, and within right parietal lobe, medially measuring 5×4 mm and slightly more laterally, measuring 2 mm. Given the multifocality and bilaterality, an embolic source was suspected. No hemorrhage, mass, midline shift, hydrocephalus, or extra-axial fluid collection was noted.

Transthoracic echocardiography (TTE) was performed, which demonstrated a 2.5×1.5 cm left atrial myxoma attached to the fossa ovalis (**Figure 1**) and a severe mitral calcification in a 1.3-cm mobile mass in the posterior chordae (**Figure 2**). The patient was started on a low-intensity heparin infusion prior to being transferred to a nearby tertiary care center for further surgical management. Continued workup at the tertiary facility with CT chest angiography demonstrated normal-appearing coronary arteries. There was no additional pathology within the chest and no pulmonary embolism. TEE again demonstrated a similar left atrial mass attached to the fossa ovalis with lobulated appearance suggestive of left atrial myxoma. Also, the calcified hypoechoic lesion measuring 1.2×1 cm on the mitral annulus posterior leaflet that was previously seen in the TTE was suggestive of extension of calcification. A second myxoma was thought to be less likely given the echogenicity

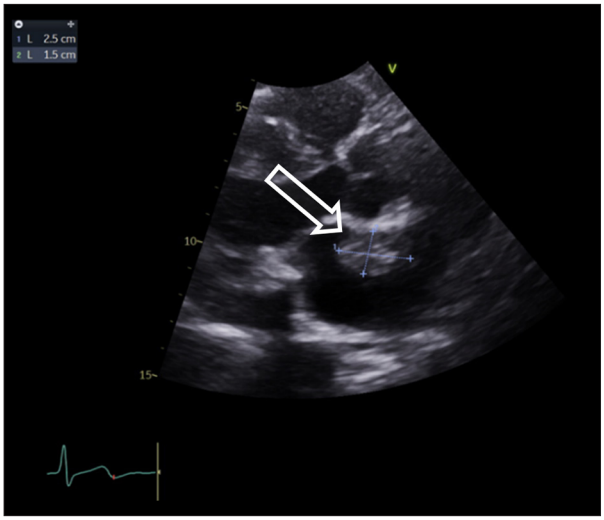

Figure 1. Transthoracic echocardiogram showing 2.5×1.5 cm fixed mass in the fossa ovalis, consistent with cardiac myxoma.

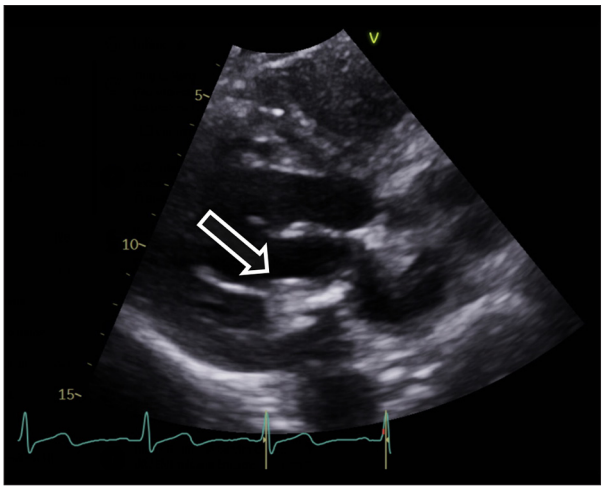

Figure 2. Transthoracic echocardiogram showing a 1.3-cm mobile mass in the posterior chordae, consistent with calcification.

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of the lesion. EF was normal at 60%, with no regional wall motion abnormalities and normal left atrial appendage without thrombus. Follow-up cardiac MRI demonstrated an approximately 2-cm left atrial mass with narrow-based attachment to the intra-atrial septum with location, morphology and cardiac MR tissue characterization consistent with left atrial myxoma. This mass was attached to the intra-atrial septum by a narrow neck. The mass demonstrated indeterminant/variable intensity on T1-weighted imaging, predominantly hypointense and isolated to hyperintense to myocardium on T2-weighted imaging. The mass also demonstrated perfusion on perfusion imaging and heterogeneous enhancement on post-contrast imaging. No intracardiac mass/thrombus was noted. Cardiovascular Surgery (CV) was consulted and recommended surgical excision. She proceeded to the operating room and underwent successful resection of her cardiac myxoma without any postoperative complications. Samples from both the calcified portion of mitral annulus and myxoma were sent for biopsy. The mitral valve sub-annular excision of calcification was evaluated with Verhoeff-Van Gieson stain, which was consistent with calcified tissue. Similarly, immunoperoxidase studies were performed using antibodies detected against calretinin and PRKAR1A showed the lesional cells to be positive for calretinin and to have retained expression of PRKAR1A, indicating a low likelihood of Carney complex.

Discussion

Cardiac myxomas are benign tumors that originate from endocardial sensory nerve tissues. They are made of myxoid extracellular matrix rich in glycoproteins and proteoglycans. While intracardiac tumors are uncommon in general, cardiac myxomas comprise 80% of cardiac tumors [1]. The most common sites for cardiac myxoma are the left atrium and the inter-atrial septum at the fossa ovalis. Its clinical manifestation may vary widely based on its size and location. Due to their location, cardiac myxomas commonly affect valvular functions, including mitral and tricuspid regurgitation, and result in left ventricular outflow tract obstruction and congestive heart failure [2,3]. Left atrial myxomas can mimic mitral stenosis and mitral regurgitation, resulting in severe enlargement of the left atrial, as observed in our patient [4]. Thus, dyspnea or respiratory distress are some of the primary symptoms secondary to significant cardiac myxoma [5]. This can be further associated with typical heart failure manifestations, including orthopnea, paroxysmal nocturnal dyspnea, pulmonary edema, cough, hemoptysis, edema, and fatigue. Additionally, cardiac myxoma can also present with arrhythmias and constitutional symptoms. While larger cardiac myxomas can cause symptoms predominantly due to mechanical interference of cardiac anatomy and flow obstruction, small cardiac myxomas with irregular tumor surfaces are at an increased risk for embolization. Tumors smaller than 3 cm are associated with a higher risk of embolic events involving cerebral infarctions [6]. Embolization of myxomatous tissues is seen in approximately 50% of patients and can affect multiple organs, including the kidneys, spleen, and central nervous system. When the central nervous system is involved, it primarily causes cerebral infarction or recurrent TIAs. Neurological manifestations of cardiac myxoma can have a range of presentations, from syncope to overt neurological deficits. Although these embolic phenomena from cardiac myxoma are common, atrial myxomas account for only 0.5% of all strokes [7]. Moreover, cerebellar involvement, as in our patient, is even rarer [8].

Embolization from cardiac myxoma mainly involves the middle cerebral artery territory, including the parietal region, temporal region, and basal ganglia. Common presenting symptoms include hypoesthesia, hemiparesis, and facial paresis [6]. Involvement of PICA territory infarcts is not as common and presents with acute vertigo, vomiting, headache, gait disturbances, and horizontal nystagmus ipsilateral to the lesion, with headache being the most common initial symptom. Cerebellar signs are a quite uncommon presentation of cardiac myxoma, and transient global amnesia may be underreported. This unusual presentation can easily deviate providers away from pursuing cardioembolic workup.

Our patient's chief concern was transient global amnesia (TGA) with associated dizziness. The differential diagnosis for TGA is broad and includes causes ranging from psychogenic and neurogenic to ischemic disease, including complex migraines, seizures, and even cardiovascular episodes. We could rule out psychogenic and neurogenic causes based on her history. We excluded some common reasons for her nonspecific dizziness, such as vertigo or presyncope, based solely on lack of positional attributes. Metabolic derangements were ruled out by initial normal laboratory work. Furthermore, our patient had no history of using illicit drugs and did not report any recent changes in her other home medications that could be a potential causative factor.

The cardiovascular origin of TGA has been questionable and is generally thought to be benign, although many authors and publications in the past have also attributed this to thromboembolic occlusion. Some of the significant cardiovascular mechanisms by which TGA has been thought to occur are transient obstruction in the territory of a posterior cerebellar artery or anterior cortical artery leading to the lack of blood supply in the deep limbic system involving memory [9]. Compared to TIA, patients with TGA without any other concerning neurological deficit have been reported to have a benign course compared to TIA regarding having a subsequent stroke and recurrent CVAs [10]. However, in the presence of other concerning neurological histories, cerebellar involvement should

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be suspected. In our patient, although her TGA lasted only for 15 minutes, it was associated with dizziness. Along with this, her history of veering toward the left side and gait instability pointed toward potential posterior circulation involvement. Transient amnesia of this nature, the predominant presentation of CVA without any focal neurological deficits, is uncommon, thus masking the concern for underlying stroke. Our patient was asymptomatic on presentation and was reluctant to consider pursuing further intervention. Without a high level of clinical suspicion, we could have easily missed her underlying stroke. Thus, it is imperative that clinicians consider the overall clinical picture and implement timely intervention to prevent adverse outcomes, including life-threatening stroke and death. Given the multifocal nature of the infarcts based on MRI, the cardioembolic source was suspected in our patient, and we thus proceeded with further cardiac imaging.

The primary imaging modality for cardiac myxomas is the echocardiogram. Myxomas present on the endocardial surface as spherical masses with speckled echogenic foci [11]. MRI provides a more detailed, accurate evaluation to narrow down the diagnosis when assessing for cardiac myxomas. These tumors have a characteristic heterogeneous appearance on MRI due to their assorted contents, including myxoid tissues, fibrous tissues, calcifications, and blood. Myxomas are also larger and more mobile than thrombi, which can be distinguishing characteristics on imaging [12]. Also, the risk of embolism of cardiac myxomas may be further complicated by calcifications. Areas of calcification are echogenic on imaging. This can be distinguished from typical cardiac myxomas, which have a heterogeneous appearance due to the different types of tissues present [11].

While cardiac CT can be used when cardiac MRI (CMR) is not readily available, CMR is still the diagnostic modality of choice in the diagnosis of cardiac myxoma. The T1 and T2 weight sequences in cardiac MRI give helpful information on the chemical microenvironment within the tumor and thus provide essential cues to differentiate the type of tumor present. Cardiac imaging alone is sufficient to decide about the need for surgery [13]. Further biopsies are obtained to establish a definitive diagnosis based on histopathology findings. Calretinin is a crucial immunohistochemical marker used to establish the diagnosis of cardiac myxoma [14]. In our patient, cardiac MRI findings showed narrow-based attachment to the intra-atrial septum, variable intensity on T1-weighted imaging, predominantly hypo-intense and isolated to hyperintense to myocardium on T2-weighted imaging, intact perfusion on perfusion imaging, and heterogeneous enhancement on post-contrast imaging. The location, morphology, and cardiac MR tissue characterization were consistent with left atrial myxoma. Additionally, immunoperoxidase studies performed using antibodies detected against calretinin and PRKAR1A showed the lesional cells to be positive for calretinin and intact PRKAR1A expression. Thus, based on CMR and biopsy findings, we concluded our patient had cardiac myxoma. It is important to emphasize that mutation in PRKAR1A is associated with hereditary conditions like Carney complex (CNC), which is an autosomal dominant disorder multiple neoplasm syndrome involving myxomas of heart, skin, neuroendocrine neoplasms, and other tissues. About 20% to 40% of the patients affected by the condition have cardiac myxoma and an associated high mortality rate. Patients with PRKAR1A mutation tend to have more severe clinical manifestations, with higher frequency of recurrent and metastatic myxoma [15].

Surgical resection is the definitive treatment for cardiac myxoma, with 2% to 5% recurrence in spontaneous cases. However, in cardiac myxomas that demonstrate familial patterns, such as patients with Carney complex, the recurrence rate is 20%. Screening is recommended in first-degree relatives of patients with recurrent cardiac myxomas after surgical removal [15]. Recurrence has been reported in cases with incomplete surgical removal of the mass; close follow-up with echocardiography is necessary to ensure adequate tumor removal.

Conclusions

In summary, this is a case of a patient who initially presented with nonspecific dizziness and transient amnesia and was discovered to have CVA involving the cerebellar region secondary to cardiac myxoma. Cardiac myxoma is well known to be associated with systemic embolization. When the CNS is involved, it often manifests as signs and symptoms related to middle cerebral artery distribution. This case illustrates an uncommon presentation of cardiac myxoma involving posterior circulation. Such an atypical presentation can mask and delay workup for underlying stoke. Early recognition of the condition is crucial to prevent potentially life-threatening consequences. An echocardiogram is a fast and noninvasive measure that should be pursued in an appropriate clinical setting when a cardioembolic source is suspected. Urgent surgical excision of the mass remains the definitive treatment. Delay in immediate intervention can result in severe thromboembolic complications. Regular long-term follow-up is necessary to monitor for recurrence. It is equally important to highlight that asymptomatic patients may consider a life-threatening condition benign if they lack an adequate understanding of the underlying pathology. Healthcare providers can play a crucial role in delivering appropriate information to help patients make informed and educated decisions.

Department and Institution Where Work Was Done

Department and institution where the work was done: Internal Medicine, MercyOne North Iowa Medical Center, Mason City, IA, USA.

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